Kawasaki disease does not affect coronaries alone: large vessels can be involved as well

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Presentation (case narrative)

A 6-month-old boy was referred with fever for 20 days, conjunctival injection, and maculopapular rash over extremities. He had normal pulses and blood pressure. Hyperemia in the Bacille Calmette–Guérin vaccination site was noted. A clinical possibility of incomplete Kawasaki disease (KD) was considered. Investigations revealed leucocytosis ($20.1 \times 10^9/L$, N : $4\text{--}11 \times 10^9/L$; neutrophils 69%); thrombocytosis ($612 \times 10^9/L$, N : $150\text{--}400 \times 10^9/L$); and raised C-reactive protein (CRP: 87 mg/L, N : < 6). Two-dimensional echocardiography showed aneurysms involving the right coronary (RCA: 9.4 mm; + 24 Z), the left anterior descending (LAD: 6.4 mm; + 16.8 Z); the left circumflex (LCx: 3.2 mm; + 6.4 Z), and the left main coronary arteries (LMCA: 3.1 mm; + 4.6 Z). Computed tomography angiography (CTA) (Fig. 1) revealed aneurysms involving all four coronaries [largest: LAD (A); and RCA (B)]; abdominal aorta (arrow in C) extending up to internal iliac (asterix in C); right (D), and left subclavian (E). He was administered intravenous immunoglobulin (2 g/kg) and infliximab (10 mg/kg), oral cyclosporine (5 mg/kg/d), methylprednisolone (30 mg/kg/d $\times$ 3 days) followed by tapering doses of oral prednisolone. Low molecular weight heparin (2 mg/kg/d),

atorvastatin (5 mg/d), and aspirin (3 mg/kg/d) were also commenced. General well-being improved and inflammatory parameters also came down. However, persistent mild elevation in CRP was noted in the follow-up for which the second dose of infliximab (10 mg/kg) was administered at the 4th month of illness. At the 8th month of follow-up, the inflammatory markers were normal and the coronary artery aneurysms partially reduced in size (LAD: 5.25 mm, + 12.1 Z; RCA: 4.3 mm, + 7.9 Z).

Discussion

Systemic artery aneurysms (SAAs) had been reported in 0.8–2.2% of patients with KD, especially in children with a delayed diagnosis who had already developed giant coronary aneurysms [1, 2]. The involvement of the aorta in SAA was considered very rare and was limited to only case reports [3–5]. Moreover, to the best of our knowledge, the involvement of the ascending aorta had never been reported before in KD [2, 3]. SAAs are usually asymptomatic (including the index child), with occasional reports of vascular insufficiency. CTA or magnetic resonance angiography is necessary to delineate the extent of involvement of systemic arteries in KD.

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Fig. 1 Computed tomography angiography imaging of index child with Kawasaki disease showing fusiform aneurysm of proximal left anterior descending artery (6.6 mm width $\times$ 16.3 mm length) (**a**); fusiform dilatation of the entire right coronary artery (proximal 8 mm, mid 5.2 mm, distal 4.2 mm) (**b**); fusiform dilatation of infrarenal

aorta (17.5 mm) (arrow in **c**) extending up to common iliac (right 12 mm, left 10.4 mm) and internal iliac arteries (right 5 mm, left 3.5 mm) (asterix in **c**); saccular aneurysm in bilateral distal subclavian arteries (right 5.6 $\times$ 10.3 mm, left 4.8 $\times$ 4.4 mm) (**d** and **e**); and dilated ascending aorta (12 mm) (**e**)

Author contribution MS and MS: writing of initial draft of the manuscript, editing and revision of manuscript at all stages of its production, and review of the literature. MS and MS are joint first authors. GA, PB, and NA: management and follow-up of the patient and editing of the manuscript. PV and SS: evaluation of the patient, follow-up of the patient, editing, and approval of the manuscript.

Declarations

Disclosures None.

Consent for publication Consent for publication was taken from parents for both content and image.

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